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A Case of Stenosis of the Conus Arteriosus Dexter.

By Dr John Hay.

The specimen which I bring before your notice to-night is one of stenosis between the conus and sinus of the right ventricle, associated with some pulmonary obstruction; the latter being caused by thickening of the pulmonary cusps, and the presence on them of vegetations.

It has seemed to me advisable in this case to depart from the usual rule; and therefore, instead of giving the clinical aspect of the case first, I will begin with a description of the *heart.*

Description of the heart, and post-mortem.

On removing the heart the right side was observed to be excessively enlarged, and to form much more than half of the whole organ. The region of the conus arteriosus dexter was noticeable as definitely enlarged, and differentiated from the rest of the right ventricle.

The whole heart weighed 15 ounces after the removal of clots, and measured $11\frac{1}{4}$ inches in circumference,—$4\frac{1}{2}$ inches being left ventricle, and the remaining $6\frac{3}{4}$ inches right ventricle, showing the comparative sizes of the two ventricles.

The apex of the heart is formed by the right ventricle.

The right auricle is much dilated and hypertrophied—its pericardial surface showing signs of fairly recent pericarditis, especially marked on the large auricular appendix.

It contained a black, soft, post-mortem clot, weighing $2\frac{3}{4}$ ounces. The average thickness of its wall is $\frac{1}{8}$ of an inch, and at the junction of the atrium with the appendix there is a thickness of $\frac{1}{4}$ inch.

The Eustachian valve is very marked, and the foramen ovale entirely occluded. The tricuspid valve admitted three fingers, and the cusps appeared normal, although perhaps they showed a slight thickening at their free margins. No dilatation of

the right ventricle; wall firm, good consistence and colour, and averages $\frac{5}{8}$ of an inch in thickness, the wall of the conus $\frac{5}{16}$ of an inch.

Separating the conus from the sinus, there is a firm muscular septum, $\frac{1}{4}$ of an inch in thickness, perforated at its centre by

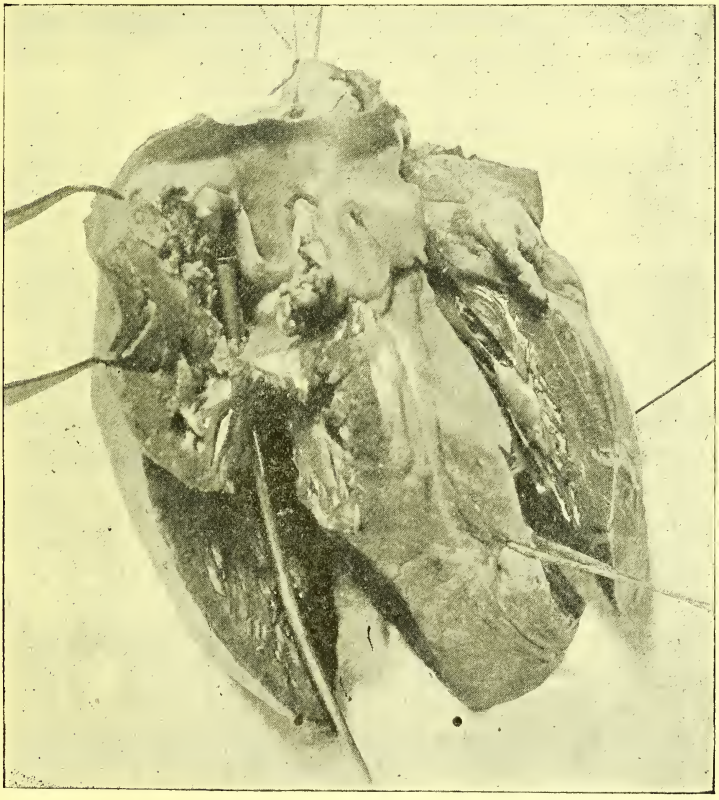

FIG. 1.—Showing septum, between conus and sinus of the right ventricle. The tube is inserted through the aperture in septum. It demonstrates also the vegetations in the conus and also the thickened pulmonary valves.

an oval aperture with a fibrous margin, the long diameter of this aperture being $\frac{1}{4}$ of an inch.

This aperture is situated $\frac{7}{8}$ of an inch below the attachment of the left anterior pulmonary cusp. The edges are fringed

with vegetations, which extend up into the conus, also on to the left anterior and posterior pulmonary cusps. The vegetations are especially numerous on the anterior wall of the conus.

Hanging from its posterior wall by a fine pedicle is a large fibrous nodulated thrombus, about the size of a marble, $\frac{1}{2}$ an inch in diameter. This would, during systole, cause a considerable increase in the obstruction to the passage of the blood past the already thickened pulmonary valves.

There are no vegetations on the pulmonary side of the cusps. The pulmonary valves are thickened and tougher than normal, and the calibre of the pulmonary artery slightly less than normal.

The ductus arteriosus was not patent.

Left auricle slightly hypertrophied. Mitral valve admitted two fingers, the cusps being normal.

The wall of the left ventricle measures, on an average, $\frac{1}{2}$ an inch ; substance firm, and good colour. The cavity is not dilated. The aortic valves competent and healthy. The infundibular portion of the left ventricle is contracted by a bulging-in of the interventricular septum ; this diminishes its size considerably, and at the base of the anterior cusp there is a marked depression, dipping in about $\frac{1}{4}$ of an inch, at the bottom of which you find the undefended space of the septum. On passing a needle through it, the needle enters the sinus of the right ventricle immediately below the septum separating it from the conus.

The origin of the aorta is placed much more directly over the right ventricle than normally, so that the blood, in escaping from the left ventricle, had to go round a corner as it were. I am inclined to think that this position of the aorta in relation to the right ventricle is congenital in origin, and analogous to the condition, though in a much less marked degree, in which the aorta arises from both ventricles, the undefended space in the interventricular septum being perforated,—this latter condition being the almost invariable accompaniment of a supernumerary septum in the right ventricle.

Post-mortem.—Some general slight œdema, more especially of legs, and skin of the abdomen.

No cyanosis.

No clubbing of the fingers or toes, but a slight increase of the curvature of the nails.

Lungs.—Adherent to the chest wall; adhesions easily broken down. Interlobar adhesions; small amount of fluid at the bases, lung substance rather pale; crepitant all over, but doughy at the bases; the bronchi rather patent and tough-walled. No signs of tubercle, past or present.

The abdomen contained about a pint of clear fluid; no signs of peritonitis.

Liver weighed 4 lbs.; was tough and nutmeg.

Spleen, 24 oz., markedly firm and fibrous; showed signs of perisplenitis.

Kidneys.—Pale on section; larger than normal; capsule stripped easily, substance tough; area of cortex normal in amount.

Pericardium.—Distended with O† of clear straw-coloured fluid. Its area corresponded with the area of dulness marked out on the chart,—this being verified by sticking long pins into the chest at points corresponding to the limits of dulness as obtained ante-mortem, and cutting down without removing the pins.

Clinical History, etc.

Mary E. Carney, 20; worked in ropeworks.

Patient admitted complaining of swelling of the legs and abdomen, also great difficulty of breathing.

Past history.—She was not blue when born, and no cyanosis was observed during childhood. When 5 years old she suffered from measles. No history of rheumatism, growing pains, chorea, or scarlet fever. It is stated that she always had a good colour, and was quite free from colds or bronchial attacks.

Present complaint dated from the age of 15. She then

received a great fright, on seeing her brother—who was injured —in the hands of policemen. She said "her heart gave a great bound," as if it had been displaced, and from that date any extra exertion or excitement produced dyspnœa and palpitation.

In the two following years, that is, until she was 17, she continued her work in the ropeworks, but at the end of that time found that the work was too heavy for her. She then stayed at home, helping with the scrubbing, etc.

She remained much the same till a month before admission, when her feet and legs began to swell.

On admission her breathing was very difficult. She presented the signs of cardiac failure and distress, œdema, dyspnœa, etc.

On 1st December I made some notes as follows: "Respirations 38, somewhat laboured. Hacking, troublesome cough, expectorating small quantity of frothy mucus.

Pulse 120; regular in time and force, small and wiry; takes 5 ounces pressure to get a pulse-tracing.

Veins of the neck greatly distended; systolic pulsation in them.

Malar capillaries dilated. On examining the chest, the whole præcordium seems to move with the systole of the heart.

The apex-beat is seen and felt in the fifth space, $4\frac{3}{4}$ of an inch from the middle line; marked epigastric pulsation is visible.

On applying the hand to the præcordium, a distinct systolic thrill is felt in the first, second, and third left intercostal spaces, extending 3 or 4 inches to the left along each space. It is rather rough in character, not a fine thrill, and, if anything, is most marked in the second space.

Cardiac dulness is greatly increased, extending $2\frac{3}{4}$ inches to the right of the middle line and 6 inches to the left, making a transverse dulness of $8\frac{3}{4}$ inch.

It is increased upwards also, there being dulness in right second space and in left first space.

On auscultation one hears a marked systolic murmur, short and harsh, all over the front of the chest; no definite first sound and no second sound to be made out anywhere. The area of maximum intensity of the murmur is at S on the chart, —the second left space just to the left of the sternum. It is here exceptionally rough, grating, and close to the ear, is also very loud in the third space, and is conducted well out to the left. There is no alteration in the murmur on getting the patient to sit up.

No systolic murmur to be heard in the neck, but there is a loud murmur in the back, loudest in the suprascapular fossæ and over the left scapula; it can be heard, though faintly, at the inferior angle of the left scapula. Nothing of note about the respiratory system, except some impaired resonance at the bases, especially the right. A few moist sounds at the left base and a considerable number at the right, associated with deficient vocal fremitus and resonance.

The liver is enormously enlarged, extending down to the umbilicus, and measuring 8 inches in the nipple line.

The spleen, hard and easily felt, 2 inches beyond the costal margin."

Five days after making the above note the patient died.

The question of diagnosis is rather interesting in this instance.

Cases of pulmonary stenosis are comparatively rare, and those of stenosis at the commencement of the conus still rarer, so that even when the physical signs are marked, one hesitates to make a positive diagnosis.

In this case pericardial effusion, considerable in extent, was an element which rendered a satisfactory understanding of the case more difficult.

The history was very little guide; in fact, it was, if anything, misleading. I may remind you that there was no history of any cyanosis, and that the patient worked up to within six weeks of her death. There was also an entire absence of any lung complications, so that practically the diagnosis depended

on physical signs,—the two most important being (1) a marked tactile thrill, systolic in time, and its area of greatest intensity being over the site of the pulmonary artery; and (2) a rough, harsh, systolic bruit, heard loudest in second and third spaces, and conducted to the left.

On these grounds, especially the combination of the two signs, I considered the case one of stenosis of the pulmonary artery.

The fact that the systolic bruit was heard almost equally loud in the third space should have given the clue to a more complete diagnosis. Dr Sansom says, in his book on *The Diagnosis of the Diseases of the Heart and Thoracic Aorta* (p. 287), when speaking of the murmur of pulmonary stenosis: "A murmur, having the characters just described, may be heard lower than the point just indicated (the second left intercostal space) in the third left space; and in such cases there may be found a stenosis (due to myocarditis) in the conus of the right ventricle, on the cardiac side of the valves, the latter being healthy."

This case also verifies the statement that dilatation of the right ventricle is rarely found with stenosis of the pulmonary artery or conus when there is no accompanying imperfection of the interventricular septum or patency of the foramen ovale. Leaving this side of the subject, let us now consider what are the causes which have given rise to the condition present in the heart under discussion; and it might be well here to notice, before mentioning the actual malformations of the conus, that there exists, as Peacock has observed, a slight normal muscular constriction between the conus and the sinus.

This is most difficult to make out in fresh specimens, because they are so flabby and soft: accordingly, I filled several hearts with 10 per cent. solution of formaline and hung them up, in this way fixing them more or less in their natural shape.

On cutting windows into them, as was done with one of the hearts on the table, one can see quite distinctly the narrowing, formed more especially by a raised muscular ridge, commencing

on the interventricular septum, and running obliquely downwards, and then round on to the anterior portion of the wall of the right ventricle.

The casts also show the same point.

I have found the constriction apparently more marked in

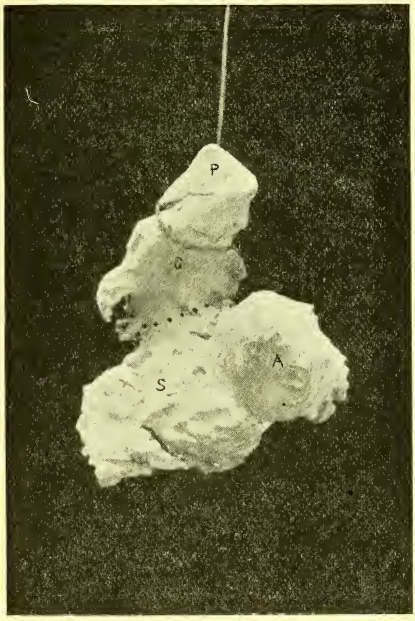

FIG. 2.—Cast of the right cavities of a heart with no valvular lesion. Patient died of phthisis. P=pulmonary artery. C=conus and S=sinus—right ventricle. A=auricle.

the hearts of phthisical patients, because there one often gets a dilatation of the conus.

However, there is normally a muscular constriction, and in some instances of a fairly marked character.

The malformations in the conus arteriosus dexter may practically be put into three classes :—

(1) A constriction between the sinus and conus.

(2) A general contraction of the conus, producing uniform narrowing.

(3) A constriction directly at or just beneath the valvular orifice of the pulmonary artery.

The second class is almost invariably due to inflammatory lesions; whilst the third is nearly always associated with a narrowing of the pulmonary artery, and shows signs of recent inflammatory mischief.

The first class is the one which concerns us more nearly to-night, and may consist of simply an exaggeration of the normal muscular constriction between the sinus and the conus. Peacock describes such a case.

More usually, however, it is caused by an inflammatory process superadded, and resulting in fibrosis, thickening, and contraction.

The opening in the septum varies greatly in size, may be multiple, and is very liable to exacerbations of inflammation at a later date.

The heart which I have brought before you belongs to this class, and is remarkable in showing no other *definite* congenital defect. I am inclined, however, to regard the dipping towards the right ventricle noticed under the anterior aortic cusp as probably congenital in origin, and pointing to a tendency to a double origin of the aorta, especially when one observes how markedly the origin of the aorta in this case lies over the upper portion of the sinus of the right ventricle.

The formation of the so-called supernumerary ventricle is generally associated with patent foramen ovale, or imperfect interventricular septum. Peacock cites ten such cases, and only two in which the stenosis between the sinus and the conus was the sole lesion,—one the child just mentioned, age 5, who presented definite signs of congenital lesion; the other, age 56, who did not.

Kussmaul reports ten other cases, and in every case the aorta sprang either from both ventricles or from the right alone.

I think, therefore, that in this case one may conclude that probably there was to begin with a congenital exaggeration of

the normal muscular constriction, formed at the junction of the sinus and conus; this constriction not being of sufficient severity to cause any marked impediment to the outflow of blood from the right ventricle, for in that case one would have expected other congenital lesions.

Later, in the conus and at the site of the muscular constriction, there probably occurred myocarditis and endocarditis. Contraction followed, producing the stenosed aperture, with its tough, fibrous margin.

During the slow evolution of these changes the right auricle and ventricle became enormously hypertrophied, in order to overcome the increasing obstruction. They were apparently equal to their task until the advent of pericarditis, which proved too much for the already handicapped right heart.

I would like here to express my indebtedness to Dr Raw for his kindness in permitting me to publish this case.